Clinics in Diagnostic Imaging (86)

P Visrutaratna, J Euathrongchit, V Kattipattanapong

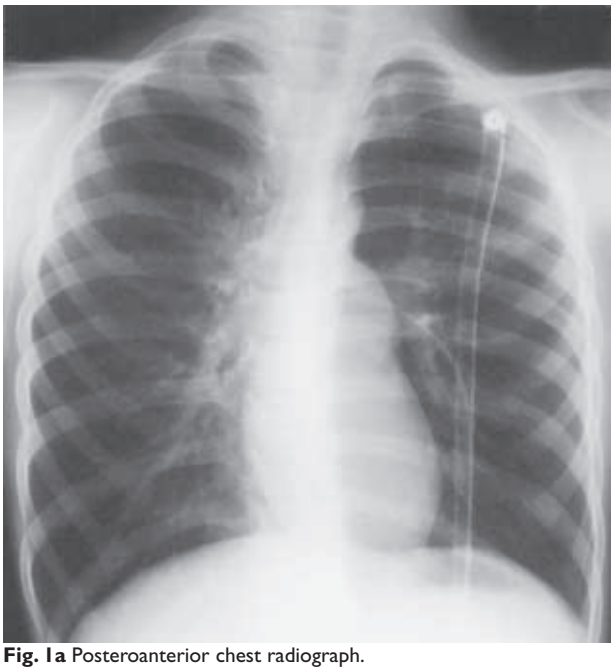

Fig. 1a Posteroanterior chest radiograph.

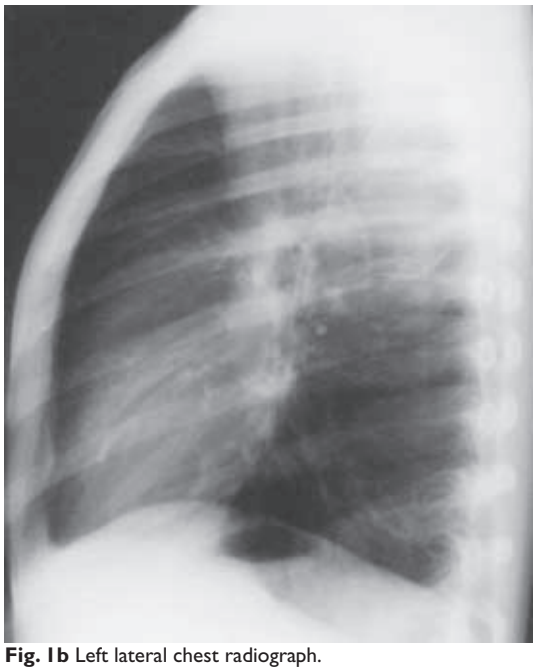

Fig. 1b Left lateral chest radiograph.

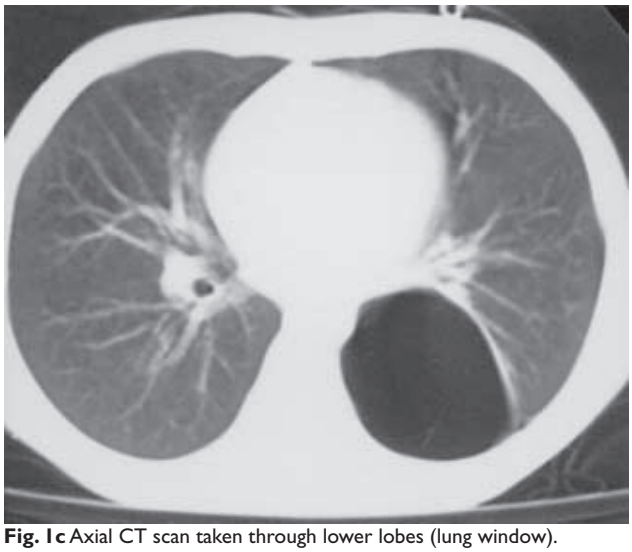

Fig. 1c Axial CT scan taken through lower lobes (lung window).

CASE PRESENTATION

An eight-year-old boy presented with a sudden onset of chest pain. He had been diagnosed to have a left-sided spontaneous pneumothorax and had a thoracostomy tube inserted at another hospital. What do his chest radiographs (Fig. 1a, b) and computed tomography (CT) of his thorax (Fig. 1c) show? What is the diagnosis?

Department of Radiology
Faculty of Medicine
Chiang Mai University
Chiang Mai 50200
Thailand

P Visrutaratna, MD
Associate Professor

J Euathrongchit, MD
Assistant Professor

Department of Surgery

V Kattipattanapong, MD
Assistant Professor

Correspondence to:
Pannee Visrutaratna
Tel: (66) 53-945450
Fax: (66) 53-217144
Email: pvisruta@mail.med.cmu.ac.th

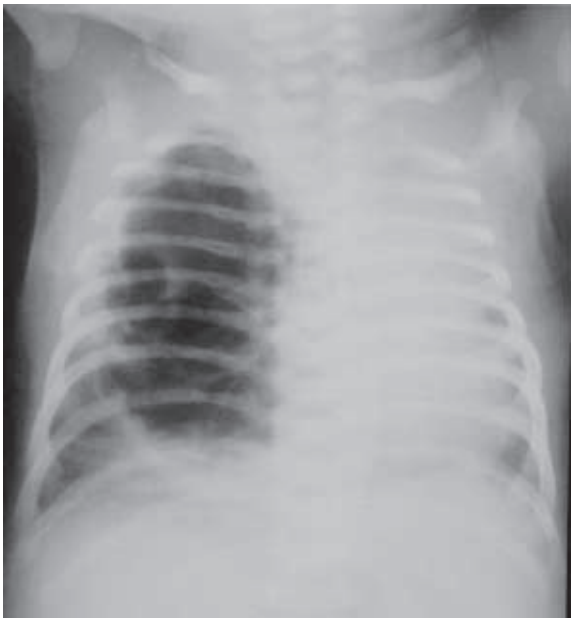

Fig. 2 Congenital cystic adenomatoid malformation. Chest radiograph shows a few large cysts and abnormal soft tissue in the right chest. Note shift of the heart and mediastium to the left.

IMAGE INTERPRETATION

The posteroanterior (Fig. 1a) and the left lateral (Fig. 1b) chest radiographs show a thin-walled cyst in the medial portion of the left lower lobe. The thoracostomy tube is visible, with its tip in the upper zone of the left lung. CT (Fig. 1c) shows that the thin-walled cyst is located in the apical segment of the lower lobe of the left lung.

DIAGNOSIS

Ruptured bronchogenic cyst.

CLINICAL COURSE

The patient had a thoracotomy, during which a ruptured cyst in the apical segment of the left lower lobe was found. The apical segment was resected. Histological examination revealed the cyst to be a bronchogenic cyst. The patient recovered and was discharged from the hospital six days after surgery. He was subsequently lost to follow up.

DISCUSSION

The causes of cystic lesions of the chest in children differ greatly from those found in adults. Congenital abnormalities are very common but malignant neoplasms are rare[1]. Intrathoracic cystic lesions in children can be classified into congenital cystic lesions and acquired cystic lesions

CONGENITAL CYSTIC LESIONS

Bronchogenic cyst, pulmonary sequestration, congenital cystic adenomatoid malformation, and congenital lobar emphysema are the four major congenital cystic thoracic lesions. Congenital diaphragmatic hernia is another important cause of intrathoracic cystic lesions in children. Chest radiography serves as the starting point for diagnosis of these cystic lesions. Sometimes, other imaging modalities such as CT, magnetic resonance (MR) imaging, ultrasonography (US), and digital subtraction angiography are needed[2]. An approach adopted by Schwartz et al is based on the initial radiographical findings. When the thoracic mass contains air, it must be examined by cross-sectional imaging, and CT is used. Conversely, when the mass is of soft tissue density and is located next to the thoracic wall, US and Doppler imaging are primarily used. CT is used only if important issues remain in doubt[3].

Bronchogenic Cyst

Bronchogenic cysts are a type of bronchopulmonary foregut malformation, which includes cystic adenomatoid malformation, bronchial atresia, sequestration, and bronchogenic cyst[4]. They may be intrapulmonary or mediastinal. Intrapulmonary cysts are due to an error in late lung bud development, and are most commonly found in the lower lobes[5]. Histologically, bronchogenic cysts are surrounded by fibrous walls containing cartilage and are lined by ciliated columnar epithelium. Most are unilocular and contain serous or mucoid fluid, unless they are infected[4]. They can life-threatening if there is compression of the tracheobronchial tree, infection, haemorrhage, or rupture[6].

Chest radiographs may show intrapulmonary bronchogenic cysts containing air, fluid, or an air-fluid level[5]. They usually have a thin sharply-defined wall, unless they are infected. They resemble pneumatocoeles radiographically, but they do not disappear spontaneously[1]. On CT, a parenchymal bronchogenic cyst appears as a well-defined mass with a smooth wall. It has an attenuation value equal to that of water, reflecting its serous fluid content. When the contents are viscous or mucoid, the attenuation value increases and may equal that of soft tissue. If communication with the airway develops, usually as a result of superimposed infection, the cyst may contain air or an air-fluid level. When complicated by infection, the cyst wall may also enhance[4].

Pulmonary Sequestration

Pulmonary sequestration is defined as a lung tissue mass that is supplied by an anomalous systemic artery (typically a branch of the thoracic or abdominal aorta), and that does not communicate with the bronchial tree via a normal bronchus[5]. This abnormality can be divided into intralobar and extralobar types, depending on the relationship to the pleura. Intralobar sequestration

is located in the visceral pleura of a lobe, usually the posterior lateral segment of the lower lobe. Extralobar sequestration occurs outside pleural boundaries but has its own pleural covering. Sequestrations large enough to cause respiratory distress in the neonatal period are usually extralobar. Intralobar sequestrations tend to be smaller and are discovered incidentally in older children and teenagers following infection[5]. On chest radiographs, lung sequestration classically appears as a triangular or oval-shaped posterior basal lung mass[7]. It may appear as cystic lung tissue containing air or air-fluid levels. Doppler US can often identify the anomalous blood supply of the lesion. Enhanced CT and MR imaging are also used to define the abnormal lung tissue and its anomalous blood supply[5].

Congenital Cystic Adenomatoid Malformation

Congenital cystic adenomatoid malformation (CCAM) is a developmental abnormality of the lung in which adenomatoid proliferation of bronchial structures results in cyst formation instead of normal alveoli[8]. There is no lobar predilection. The lesions are limited to a single lobe in over 95% of cases[8]. They communicate with the tracheobronchial tree and usually receive their blood from the pulmonary circulation. CCAM is usually discovered in neonates because of respiratory distress but may occasionally be discovered in older children or adults who have recurrent infection. There have been reports of malignancies such as rhabdomyosarcoma developing in CCAM[5].

CCAM is divided into three types based on the size of the cysts contained in the lesion and other histologic criteria. Type 1 lesions, which make up 50% of cases, have one or more large cysts (2-10 cm). Type 2 lesions, which account for 40% of cases, consist of smaller multiple cysts (0.5-2 cm). Type 3 lesions, which account for 10% of cases, appear solid on gross examination but contain multiple 0.3 to 0.5 cm cysts on microscopical examination[8]. On chest radiographs and CT, the findings vary according to the type of malformation[8,9]. The number, size, and amount of fluid within the cysts determine whether the lesion appears predominantly cystic or solid on chest radiographs[8]. The cystic components become air-filled within hours to days after birth (Fig. 2). In severe abnormalities, the lung may be hyperexpanded with mediastinal shift and herniation of the lung to the contralateral side[5]. On CT, visualisation of cystic changes associated with abnormal soft tissue helps differentiate CCAM from other causes of abnormal radiolucency in neonates, such as congenital lobar emphysema and pulmonary interstitial emphysema[8].

Congenital Lobar Emphysema

Congenital lobar emphysema is a congenital overexpansion of a pulmonary lobe that may be the result of an abnormality related to the airway. Weakness in the airway wall or intrinsic or extrinsic compression of the airway is thought to lead to overdistension and air trapping in alveolar spaces[5]. Focal areas of bronchomalacia, kinks, webs, mucosal folds, and crossing vessels have all been implicated in its pathogenesis[3]. In about one-half of the cases, the cause is unknown[5]. Congenital lobar emphysema with an increased number of alveoli, i.e. polyalveolar lobe, is an uncommon variant[3]. Congenital lobar emphysema is seen more commonly in males. It occurs also most commonly in the left upper lobe, followed by the right middle lobe and the right upper lobe. Occasionally, it affects more than one lobe. The lower lobes can also be involved but this site is rare.

Patients with this abnormality usually present with respiratory distress in the perinatal period. Older infants can present with either respiratory distress or repeated infections[10]. The initial chest radiographs taken soon after birth frequently show radiopacity of the involved lobe. Subsequent radiographs show increasing radiolucency as the foetal lung fluid is replaced by air[10] (Fig. 3a). Compression of the ipsilateral lung, mediastinal shift, and compression of the contralateral lung are sometimes seen. Chest radiographs are usually sufficient to diagnose congenital lobar emphysema, but pneumothorax and CCAM sometimes have similar radiographical and clinical features. CT is helpful in cases where the diagnosis is in doubt[5]. It reveals the overdistended lobe with stretched and attenuated vascular structures, which extends out to the lung peripherphy (Fig. 3b). As on chest radiographs, there are adjacent compressive atelectasis and contralateral mediastinal shift[4].

Congenital Diaphragmatic Hernia

Herniation of the abdominal contents can occur through various portions of the diaphragm, but it most often occurs through the posterolateral developmental defect (foramen of Bochdalek). Left-sided hernias are more common. The symptoms depend on the degree of herniation. Small hernias sometimes cause no herniation while larger ones produce immediate and severe respiratory distress[7]. On chest radiographs, the fluid-filled loops of bowel are seen initially as a water-density mass in the left hemithorax. The mass is located along the inferior aspect of the chest, with varying degrees of contralateral mediastinal shift[10]. Intrathoracic bowel can sometimes mimic a cystic

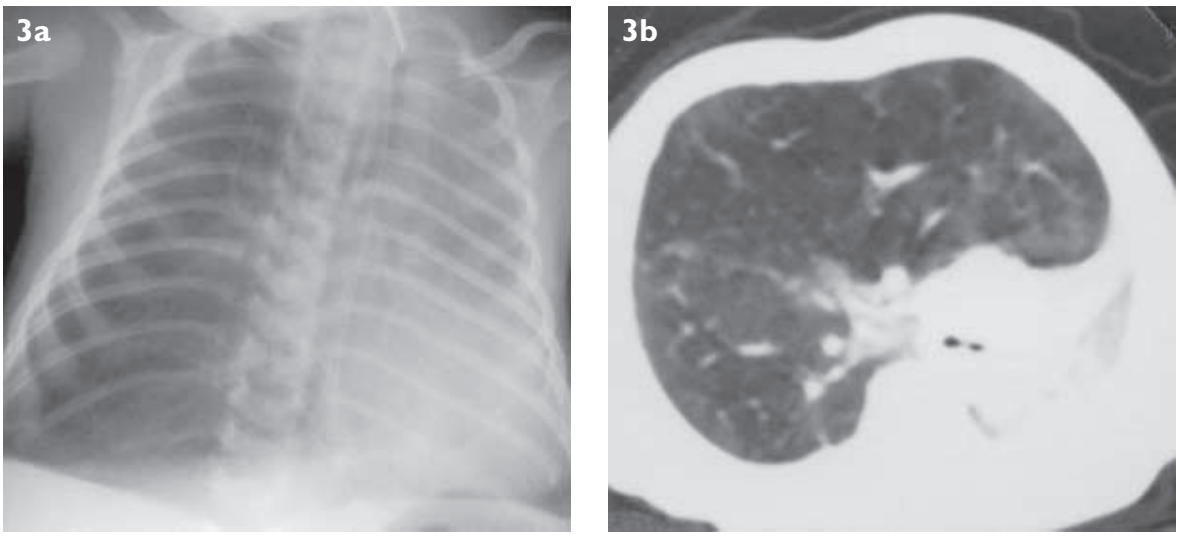

Fig. 3 Congenital lobar emphysema. a) Chest radiograph shows hyperaeration of the right lung, herniation of the right lung to the left, and shift of the mediastinum to the left. b) CT scan taken through the carina (lung window) shows an overdistended right upper lobe with stretched and attenuated vascular structures and herniation of the right upper lobe to the left.

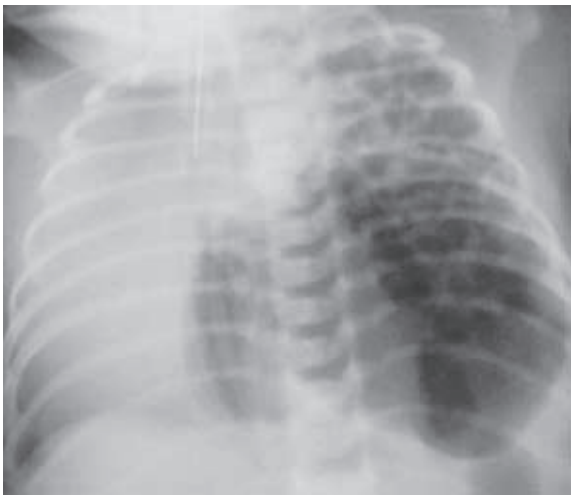

Fig. 4 Congenital diaphragmatic hernia. Chest radiograph shows the herniated stomach and bowel loops in the left chest. The heart and mediastinum are shifted to the right.

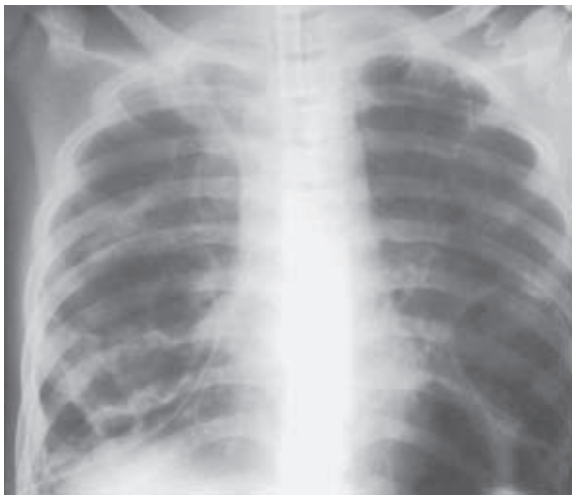

Fig. 5 Pneumatocoeles in child with staphylococcal pneumonia. Chest radiograph shows multiple thin-walled cysts in both lungs. There is a thoracostomy tube in the right chest because of pneumothorax from a ruptured pneumatocoele.

lung mass (Fig. 4), but loss of definition of the hemidiaphragm, lack of bowel in the abdomen, and abnormal position of support lines and tubes are usually diagnostic in the neonatal period[5].

ACQUIRED CYSTIC LESIONS

Inflammatory Pneumatocoele

Inflammatory pneumatocoeles result from a rapidly-progressive inflammation in which there are plugging of the smaller bronchi in the affected areas, destruction of the distal alveolar tissue, and cystic hyperexpansion of the air space[10,11]. Pneumatocoeles are found much more often in children than in adults. They are seen particularly in infants and children with staphylococcal pneumonia[11]. However, they have also been described in pneumonia due to other micro-organisms such as Streptococcus pneumoniae, Klebsiella, Hemophilus influenzae, and in measles pneumonitis[10].

On chest radiographs, they appear as thin-walled, rounded radiolucencies in areas of pneumonic consolidation. They can be single or multiple (Fig. 5). As the surrounding consolidation resolves, the pneumatocoeles become more apparent and/or enlarge. Thus, they are usually seen during the resolution stage of pneumonitis[10]. In some cases, they can become large enough to cause a tension phenomenon. Occasionally, they rupture and cause a pneumothorax. Most often, however, they resolve slowly and disappear within two to three weeks[10].

Traumatic Pneumatocoele

Traumatic pneumatocoeles infrequently accompany blunt chest trauma and pulmonary contusion. They are believed to result from tears within the pulmonary

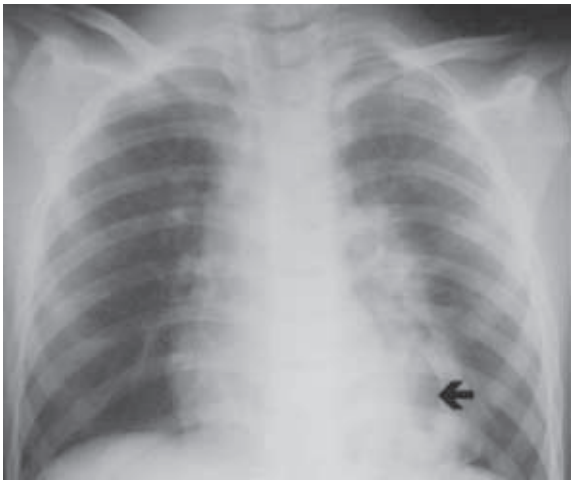

Fig. 6 Traumatic pneumatocoele. Chest radiograph shows opacification of the medial portion of the left chest from pulmonary contusion. Note cavity due to traumatic pneumatocoele (arrow).

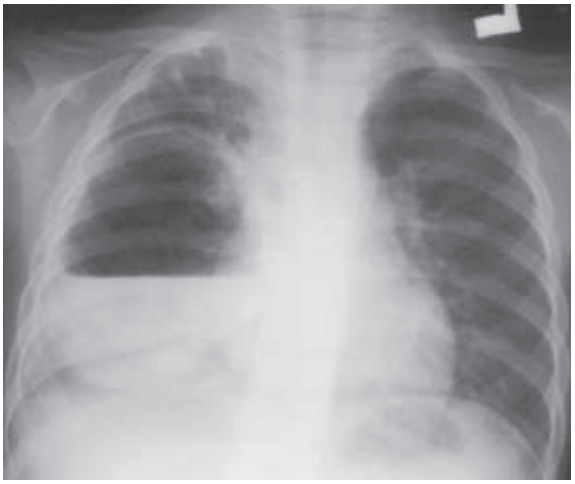

Fig. 7 Lung abscess. Chest radiograph shows a moderately thick-walled cavity with air-fluid level in right lower lobe. Note minimal infiltration adjacent to the abscess.

parenchyma caused by sudden elastic recoil after forceful and rapid compression. They are much more common in children and young adults, perhaps as a result of the proportionally greater elasticity of the chest wall of younger people[10]. Traumatic pneumatocoeles usually present within 12 to 24 hours of injury, although they may not appear on chest radiographs until the enveloping pulmonary contusion begins to dissipate 24 to 72 hours after injury (Fig. 6). On CT, they appear as uni- or multilocular cavities, with or without air-fluid levels[4]. Traumatic pneumatocoeles slowly become smaller, disappearing after two to three weeks.

Pulmonary Abscess

Pulmonary abscesses are necrotic cavities filled with purulent material, which may have air-fluid levels if they have eroded into airways. They are usually caused by aspiration of anaerobic oral flora but can be caused by other organisms such as Streptococcus and Klebsiella[10]. On chest radiographs, air-fluid levels are frequently present. The abscess walls are hazy, and vary in thickness (Fig. 7). Haziness, due to surrounding inflammation, distinguishes the abscess from a pneumatocoele or a bronchogenic cyst in most cases[12]. On CT, pulmonary abscesses appear as spherical hypodense masses with thick irregular walls, especially along their inner margins. They have a poorly-defined external surface because of surrounding infected parenchyma[4].

Pulmonary Cysts in Systemic Disease

There are many systemic disorders that may produce cystic changes in the lungs, such as hyper immunoglobulin E syndrome, hydatid cysts, Langerhans cell histiocytosis, Marfan syndrome, proteus syndrome, and neurofibromatosis[10].

ABSTRACT

An eight-year-old boy presented with a sudden onset of chest pain. He had been diagnosed to have a left spontaneous pneumothorax. Chest radiographs and computed tomography of the chest showed a thin-walled cyst in the left lower lobe. Thoracotomy and a segmentectomy of the apical segment of the lower lobe was performed, confirming the diagnosis of a ruptured bronchogenic cyst. Imaging findings of various pulmonary cystic lesions in children are discussed.

Keywords: chest, children, bronchogenic cyst, cystic adenomatoid malformation, congenital lobar emphysema.

Singapore Med J 2003 Vol 44(6):325-329

REFERENCES

1. Mercado-Deane MG, John SD. Acute presentation of cystic and mass-like lesions of the chest in children. Emerg Radiol 2001; 8:75-84.
2. Takeda S, Miyoshi S, Inoue M, Omori K, Okumura M, Yoon HE, et al. Clinical spectrum of congenital cystic disease of the lung in children. Eur J Cardiothorac Surg 1999; 15:11-7.
3. Schwartz DS, Reyes-Mugica M, Keller MS. Imaging of surgical diseases of the newborn chest. Intrapleural mass lesions. Radiol Clin North Am 1999; 37:1067-78.
4. Siegel MJ. Pediatric Body CT. Philadelphia: Lippincott Williams & Wilkins, 1999.
5. Williams HJ, Johnson KJ. Imaging of congenital cystic lung lesions. Paediatr Respir Rev 2002; 3:120-7.
6. Ribet ME, Copin MC, Gosselin BH. Bronchogenic cysts of the lung. Ann Thorac Surg 1996; 61:1636-40.
7. Swischuk LE. Imaging of the Newborn, Infant, and Young Child. 4th ed. Philadelphia: Lippincott Williams & Wilkins, 1997.
8. Donnelly LF, Frush DP. Localized radiolucent chest lesions in neonates: causes and differentiation. Am J Roentgenol 1999; 172:1651-8.
9. Kim WS, Lee KS, Kim IO, Suh YL, Im JG, Yeon KM, et al. Congenital cystic adenomatoid malformation of the lung: CT-pathologic correlation. Am J Roentgenol 1997; 168:47-53.
10. Hernanz-Schulman M. Cysts and cystlike lesions of the lung. Radiol Clin North Am 1993; 31:631-49.
11. Shamji FM, Sachs HJ, Perkins DG. Cystic disease of the lungs. Surg Clin North Am 1988; 68:581-620.
12. Swischuk LE, John SD. Differential Diagnosis in Pediatric Radiology. 2nd ed. Baltimore: Williams & Wilkins, 1995.